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## Case Report

# Linear scleroderma en coup de sabre presenting with seizures <sup>☆</sup>

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### ABSTRACT

Scleroderma is a rare connective tissue disorder categorized into systemic sclerosis and localized scleroderma, also called morphea. Linear scleroderma of the scalp, also called en coup de sabre, is infrequently associated with neurologic symptoms. We describe a case of linear scleroderma en coup de sabre in a 28-year-old female presenting with seizures and characteristic cutaneous lesions. Imaging findings over a course of 7 years demonstrated waxing and waning signal changes. MR perfusion and spectroscopic imaging, demonstrating decreased cerebral blood volume, increased mean transit time, and decreased metabolites, was performed during a time of progressing radiological and clinical findings. Comparison with other reports in the literature supported several clinical and imaging findings that while not pathognomic, highly suggest the diagnosis of linear scleroderma en coup de sabre. Hyperintense signal on T2W magnetic resonance imaging and contrast enhancement on computed tomography and magnetic resonance imaging have been the most commonly described imaging findings. To our knowledge, no previous description of spectroscopic or perfusion imaging of linear scleroderma en coup de sabre have been reported. It is our hope that this report may add MRS and magnetic resonance perfusion findings to a growing knowledge of this rare entity.

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## Introduction

Scleroderma is a rare connective tissue disorder that is categorized as systemic sclerosis or localized scleroderma, the latter also called morphea. Unlike systemic sclerosis, localized scleroderma is not associated with Raynaud phenomenon, sclerodactyly, erythema, hand edema, or nailbed capillary changes [1]. Localized scleroderma presents in 5 ways: circumscribed,

generalized, linear, mixed, and pan-sclerotic [1]. Linear scleroderma usually manifests as cutaneous linear scar-like lesions of the head or limbs [1]. It is the most frequent form of scleroderma encountered in children [2]. If there is involvement of the head, the linear scleroderma is referred to as en coup de sabre (LScs) [3]. This is the most frequent form of morphea of the scalp. Previously, it was believed that linear scleroderma and its variants were limited to skin involvement, with multi-system involvement being exclusive to systemic scleroderma.

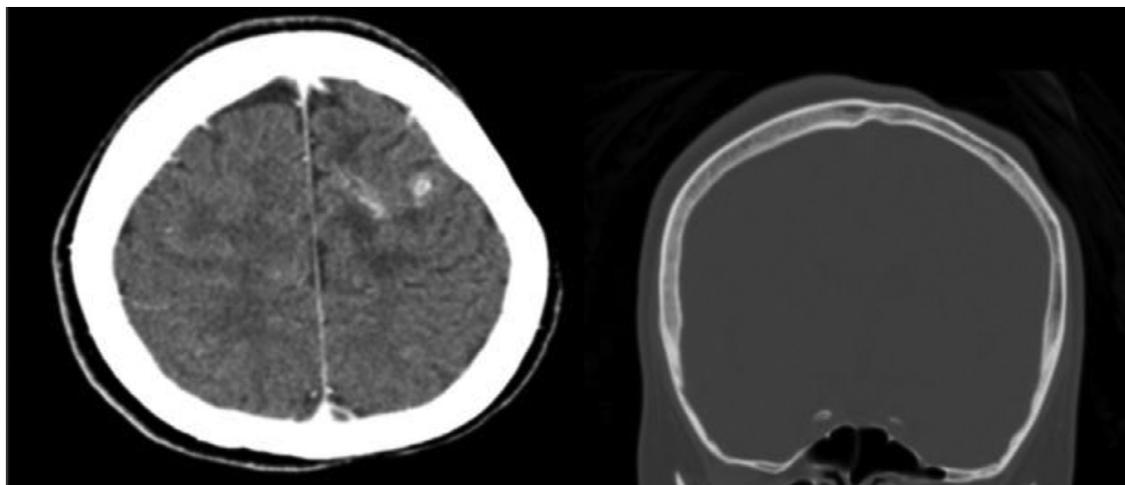
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**Fig. 1 – (Left) Axial contrast-enhanced CT of the head at the time of ED presentation demonstrating enhancing linear and nodular lesions within the left frontal lobe at the gray white junction with surrounding gliosis/edema. Notice the subtle left frontal lobe volume loss. (Right) Axial CT of the head with bone algorithm demonstrating scalp thinning and subtle thinning of the left parietal calvarium.**

However, this has been shown to not be the case as rheumatologic, ophthalmologic, and neurologic symptoms have been noted in up to 20% of cases, with the nervous system being the most common extracutaneous system involved [4]. In 1990, a patient with LScs experiencing seizures and hemiparesis was noted to have brain lesions ipsilateral to characteristic cutaneous lesions [5]. Since then, other cases of cortical and subcortical LScs lesions have been reported ipsilateral to the skin findings. Histological studies document band-like sclerosis of the leptomeninges, low grade vasculitis, and focal cerebral necrosis, suggesting LScs to be an inflammatory process [6,7]. Progressive facial hemiatrophy, also known as Parry-Romberg syndrome may be related to LScs and is also characterized by unilateral atrophy of the skin, subcutaneous tissue, and bony involvement. Neurologic complications associated with Parry-Romberg syndrome generally involve the trigeminal nerve branches [8,9,15]. As localized scleroderma is still not well understood and extracutaneous involvement of disease in LScs uncommon, this case aims to provide additional imaging perspective into this rare diagnosis.

### Case report

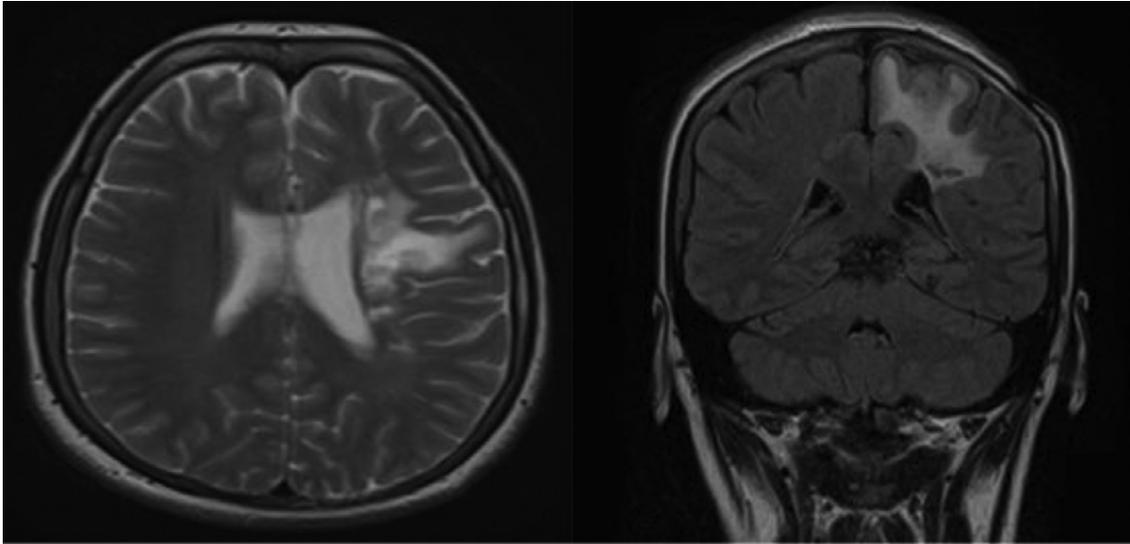
A 28-year-old woman presented to the Emergency Department for evaluation of chronic right upper and lower extremity weakness. She developed seizures at the age of 21. At that time, magnetic resonance imaging (MRI) was performed in Mongolia demonstrating a brain lesion, interpreted as a possible abscess. She was treated with antibiotics and her condition improved. She was also started on carbamazepine for management of her seizures. One year prior to presenting to our institution, seizure activity had increased in frequency to 5-6 times a month. The seizures manifested as a tingling sensation in the extremities followed by diffuse convulsions, without loss of consciousness. In addition, she began suffering dif-

fuse headaches with worsening right upper and lower extremity weakness, leading to balance and gait problems. Outside of the neurologic symptoms, the patient did not note any constitutional, gastrointestinal, respiratory or infectious symptoms. Neurologic evaluation revealed right upper extremity rigidity and decreased strength in the right upper and lower extremities as well as increased right sided reflexes.

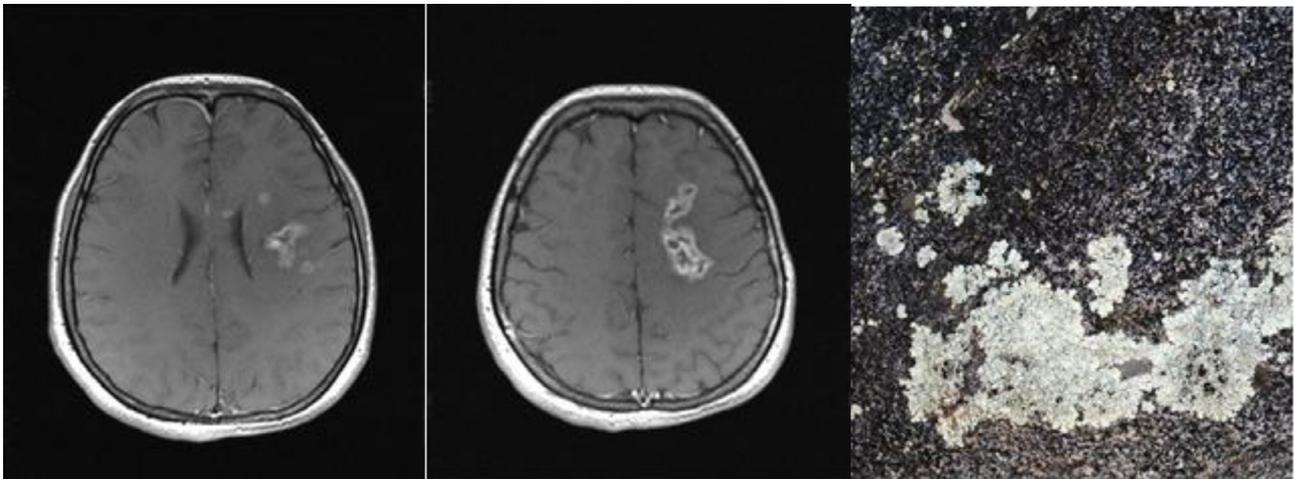
Computed tomography (CT) of the head performed in the Emergency Department is seen in Fig. 1 with select axial and coronal contrast-enhanced head CT images demonstrating left parietal scalp thinning. There was also subtle thinning of the left parietal diploic space. The CT head also showed nodular and linear enhancing lesions at the gray white matter interface with surrounding vasogenic edema in the left superior frontal gyrus.

A follow up brain MRI can be seen in Fig. 2 and the center image of Fig. 7 which demonstrated hyperenhancement of the left corona radiata, left centrum semiovale, and left superior frontal gyrus branching to the gray white matter interface. There was no diffusion restriction. A few foci of gradient-recalled echo (GRE) signal dropout were noted within the left centrum semiovale, left midbrain, and left cerebellar hemisphere.

Fig. 3 displays images from an MRI obtained 7 years earlier. Confluent serpiginous enhancement in the centrum semiovale branch into the peripheral white matter of the left superior frontal and precentral gyri. The central nonenhancement was noted with a few areas demonstrating frank cavitation on follow-up imaging. Fig. 3 depicts the more confluent areas of enhancement while Fig. 7 highlights the projections into the peripheral white matter. Fig. 4 demonstrates MR spectroscopy performed over the site of abnormality at the time of presentation prior to methotrexate treatment, depicting a choline peak of 4.6, creatinine peak of 4.2, and N-acetylaspartate (NAA) peak of 5.0. Normal brain parenchyma demonstrated a choline peak of 5.1, creatinine peak of 6.6, and NAA peak of 12.2. Spectroscopy performed



**Fig. 2 – (Left) Axial T2W MRI of the brain at time of ED presentation demonstrating hyperintense lesion within the left corona radiata. (Right) Coronal FLAIR MRI of the brain demonstrating hyperintensity in the left centrum semiovale and corona radiata.**

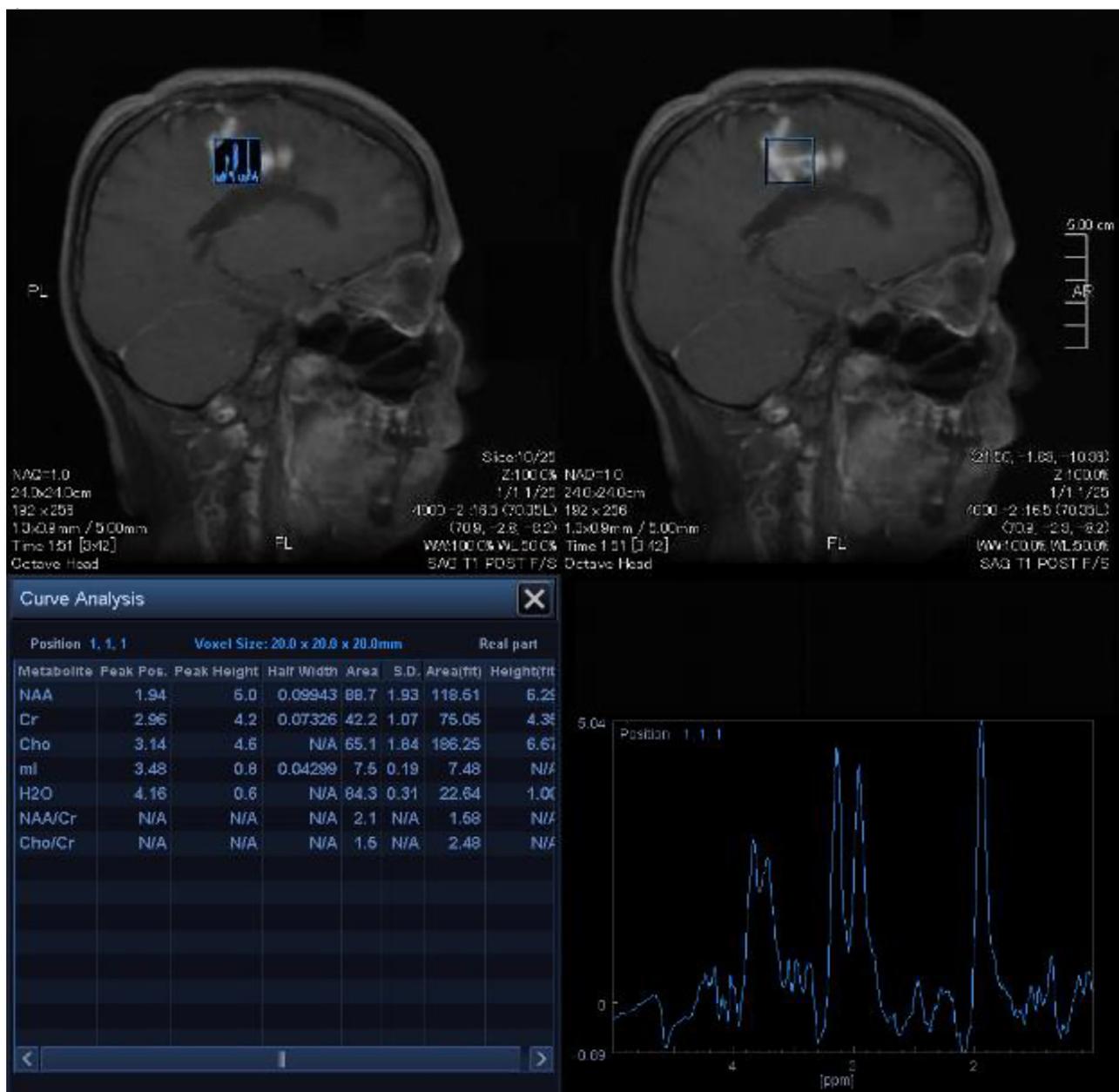


**Fig. 3 – (Left) Axial T1W postcontrast MRI of the brain obtained 7 years prior to presentation demonstrating multifocal enhancing lesions extending into the corpus callosum and precentral gyrus. (Middle) Axial T1W postcontrast MRI of the brain demonstrating “lichenoid” branching enhancement with central necrosis and marginal enhancement. (Right) Lichen growing on rock. (Image copyright licensed and used with permission courtesy of shutterstock.com.)**

over the site of abnormality demonstrated slightly decreased choline and creatinine peaks compared to the contralateral normal brain as well as a significant decreased NAA. MRI perfusion in Fig. 5 revealed relative decreased cerebral blood flow, cerebral blood volume, and no major alteration of the mean transit time although, there was very slight prolongation of the mean transit time in the left centrum semiovale.

Workup with immunologic and inflammatory serologic markers were all negative. Subsequently, the patient followed up in the dermatology clinic. The hyperpigmented sclerotic scalp lesions were noted to be characteristic for localized scleroderma as seen in Fig. 6. Dermatology documented a linear “indentation/deformity” with hyperpigmentation along

the left temple and upper back over the thoracic spine. Per the patient, this cutaneous finding developed at age 12. These dermatological findings then prompted reevaluation of imaging findings. Fig. 7 demonstrates the change in appearance of the lesion on T1-weighted contrast enhanced MRI at 3 separate time points: 7 years prior to presentation, at the time or presentation and 5 months later. Initial imaging 7 years prior showed extensive T2 FLAIR high signal and gyral expansion involving the left superior frontal gyrus, precentral gyrus, and paracentral lobule. “Lichenoid” branching enhancement extends from the left centrum semiovale into the adjacent gyri and corpus callosum. On this initial exam, there was no evidence for brain volume loss. Thinning of the ipsilateral



**Fig. 4 – MR spectroscopy performed over the site of abnormality prior to methotrexate treatment demonstrates a choline peak of 4.6, creatinine peak of 4.2, and NAA peak of 5.0 (normal brain parenchyma: choline peak of 5.1, creatinine peak of 6.6, and NAA peak of 12.2).**

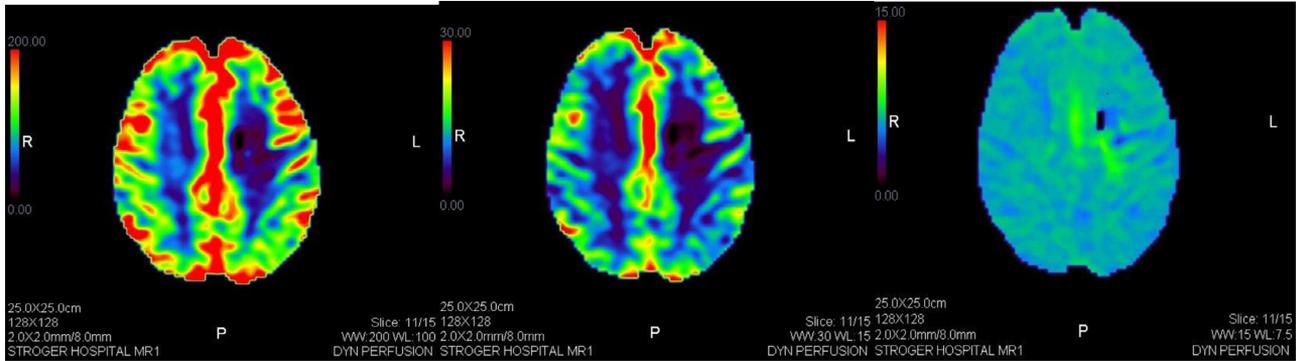
calvarium and the overlying soft tissues was present on this initial imaging study. At the time of the patient's presentation to the ED, MRI showed progression of confluent T2 high signal hyperintensity in the same region of white matter with new swelling of the precentral and postcentral gyri. Similar lichenoid enhancement was noted with resolution of the anterior centrum semiovale enhancement. Enhancement was no longer identified within the left aspect of the corpus callosum, but new foci of enhancement were noted in the left frontal lobe.

Once other infectious etiologies were ruled out, the patient was subsequently treated with methotrexate and prednisone alongside her longstanding carbamazepine with some improvement of her symptoms. A follow up MRI 5 months

later after 3 months of methotrexate therapy is seen in the right image of Fig. 7, demonstrating waning enhancement in the left centrum semiovale and corona radiata. The combination of dermatologic findings with neurologic symptoms and ipsilateral brain MRI findings supported the diagnosis of linear scleroderma en coup de sabre.

## Discussion

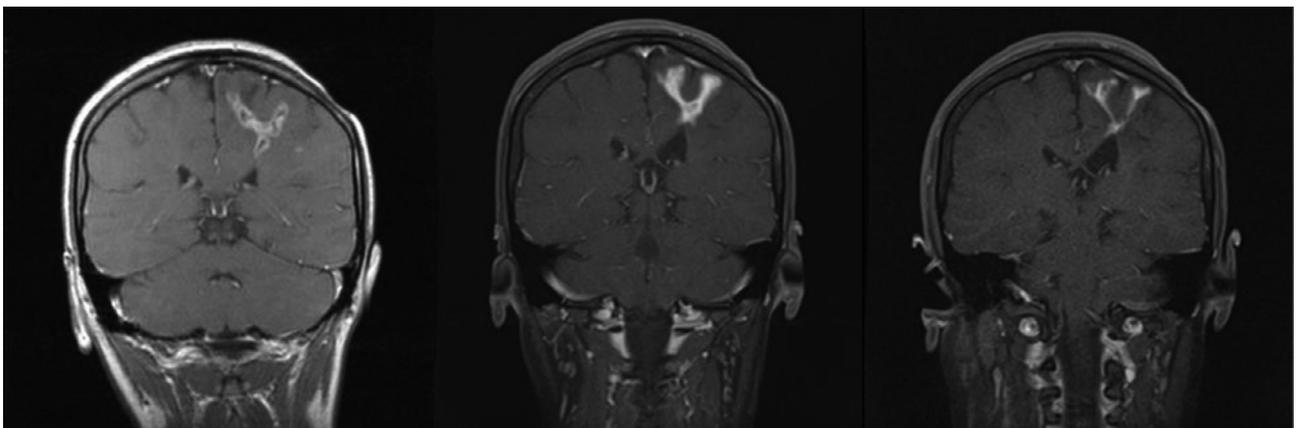
Linear scleroderma en coup de sabre is a rare form of localized scleroderma. LSCs remains a clinical diagnosis, with no clear serologic, imaging or pathologic features considered pathog-



**Fig. 5** – MR perfusion imaging was performed shortly around the time of ED presentation. (Left) Cerebral blood flow and (Center) cerebral blood volume are relatively decreased in the left centrum semiovale compared to the contralateral side. (Right) Minimal increased mean transit time in the left centrum semiovale.



**Fig. 6** – (Left) Photograph demonstrating hyperpigmented and sclerotic cutaneous band along the left forehead. (Center) Photograph demonstrating extent of the cutaneous lesion to the scalp area. (RIGHT) 3-D reconstruction from the patient's MRI demonstrating a furrow along the left scalp corresponding with the cutaneous lesion seen on exam.



**Fig. 7** – Coronal T1 postcontrast MRI images demonstrating the waxing and waning intra-axial lesions. (LEFT) Initial imaging from 7 years prior to presentation demonstrates branching centrally necrotic enhancing lesions in left centrum semiovale extending to the left superior frontal and middle frontal gyri. There is no evidence for brain volume loss. Note the ipsilateral thinning of the calvarium and scalp. (Center) Imaging from 7 years later demonstrates interval progression in the degree of enhancement accompanied with new ex vacuo dilation of the body of the left lateral ventricle. (Right) Imaging 5 months after the CENTER MRI and 3 months after methotrexate treatment demonstrates decreased but persistent enhancement with residual ex vacuo dilation of the left lateral ventricle.

nomonic. More prevalent in women, the incidence is highest at menarche. The average age of onset is 13 years of age [3]. Our patient noticed cutaneous manifestations at age 12. As with our patient, complex partial seizures are the most common neurologic complication of scleroderma [3].

Scl-70 is often utilized as a serologic marker for systemic sclerosis, but serologic markers for localized scleroderma are variable [3]. Less frequently reported, histological descriptions of LSCs have described leptomeningeal band-like sclerosis, intraparenchymal calcifications, ectatic vessels, gliosis, chronic inflammation, and perivascular infiltrates [6,10,7]. Treatment remains controversial and no clear regimen or algorithm has been established

CT of the head in Fig. 1 shows the cortex of the left parietal bone was relatively symmetric to the right, without sclerotic or lucent changes. Frank bone deformities were not seen in our case but have been described [11]. Like other case reports, the radiological central nervous system (CNS) manifestations were noted ipsilateral to the cutaneous lesion. Clustered nodular and curvilinear enhancing lesions extended to the peripheral white matter of left superior frontal and precentral gyri from the centrum semiovale. While comparison to a prior exam suggested new gyral expansion and vasogenic edema, there was also unequivocal evidence for underlying brain volume loss with parasagittal frontal lobe sulcal widening and left lateral ventricle ex vacuo dilation [12]. No calcifications were noted but have been described in other case reports [13,12].

Most of the lesions appeared to be confluent but discontinuous subcentimeter lesions were also scattered in the left cingulate gyrus and anterior body of the corpus callosum. The enhancement pattern reminded the author of lichen with patchy islands of growth seemingly following the scaffolding of regional white matter tracts. Patient's age and long-standing findings argued against a multifocal high-grade glioma and other malignant processes. Unilateral multifocal lesions all demonstrating peculiar branching enhancement were not suggestive of a demyelinating process. Primary central nervous system (PCNS) vasculitis or conditions like Behcet's disease were considered along with chronic fungal or granulomatous infections. On baseline MRI, the T2 prolongation was related to vasogenic edema; however, on the follow up MRI, Fig. 3 the confluent T2 hyperintensity within the left centrum semiovale and corona radiata appears to reflect a superimposition of edema on regional areas of gliosis.

Information on the prevalence and exact imaging findings in LSCs has not been well described, as imaging is usually only done for symptomatic patients [3]. It is likely that even among asymptomatic LSCs patients, MRI would likely reveal at least some characteristic findings [13]. Reports of radiologic findings in Progressive facial hemiatrophy have also demonstrated hyperintense T2 lesions [14]. The first pathologic finding of LSCs seen in the literature was by Chung in 1995, which showed leptomeningeal sclerosis with intraparenchymal calcifications and anomalous ectatic vessels. The earliest imaging findings were reported in 2001 by Stone with the MRI demonstrating mild focal atrophy on T1-weighted imaging, gadolinium enhancement and hyperintense foci in the gray and white matter of the right hemisphere with pathology showing lymphocytic and monocytic infiltration with area of foamy macrophages and zone of necrosis interpreted as low-

grade vasculitis with associated focal cerebral necrosis. In this case, repeat MRI in 6 months demonstrated partial resolution of the previously seen T2 hyperintense lesions in the right hemisphere with a new right frontal lobe T2 hyperintense lesion. A second repeat MRI demonstrated a new T2 hyperintense enhancing lesion in the left thalamus [7]. This case illustrates the waxing and waning nature of LSCs. In 2004, Appenzeller presented a case of LSCs which reaffirmed T2 and FLAIR hyperintense lesions as a likely defining characteristic. Atrophy, calcifications and thinning of the skull ipsilateral to skin lesions was also seen on CT [11]. Holland reported a case in 2006 of a patient with LSCs that demonstrated left frontal lobe calcifications on head CT and T2 hyperintense signal lesions on MRI with pathologic findings of chronic perivascular lymphocytic inflammation with aggregates of dilated vessels and partially organized thrombi interpreted as a primary vasculitis. PET CT was performed in this case, which was not reported previously in the literature, demonstrating nonspecific hypometabolic areas in the same region [12]. A case from 2018 written by Duman demonstrated multiple T2 hyperintense lesions with associated scalp thinning and thinning of the corresponding frontal and parietal bones with mild contrast enhancement. The most recent case from 2019, reported by Magro showed multiple enhancing lesions in the bilateral juxtacortical, subcortical, and periventricular white matter and in the body of the corpus callosum with soft tissue and bone defects in the left parietal bone on contrast enhanced T1-weighted imaging [16]. Corresponding pathology showed cortical necrosis with lymphocytes surrounding capillaries with some vascular thrombosis interpreted as lymphocytic vasculitis. Through these case reports, commonalities become apparent with the case presented. Decreased MR perfusion seems to correlate with prior PET hypometabolism. MR perfusion deficits have been described in cases of CNS vasculitis and would correlate with the pathological findings of a primary vasculitis.

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## Conclusion

While there are no pathognomonic imaging findings, case reports have described certain recurring findings on CT and MRI. MRI commonly demonstrates focal atrophy, ipsilateral blurring of the gray-white matter junction, T2 and FLAIR hyperintense lesions, and postcontrast enhancing lesions. MRI findings in the literature are summarized in the previous discussion, with the mainstay radiologic findings showing T2 hyperintense lesions as the most reported finding. Pathologic specimens seem to point to vasculitis as an underlying etiology. Linear scleroderma en coup de sabre remains a rare diagnosis, but a constellation of imaging findings in conjunction with a dermatological exam may permit this as a reasonable differential diagnosis. The lesions in this case do have a somewhat infiltrative appearance to them and may raise concern for a high-grade primary CNS lesion in someone presenting at a later age. MR perfusion and spectroscopy could potentially be reassuring to exclude the possibility of an infiltrating high-grade glioma. To our knowledge no one has reported the MR perfusion and spectroscopic findings associated with LSCs.

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